

VISUAL IMPAIRMENT IN NASOPHARYNGEAL CARCINOMA

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Abstract

Introduction: The most common complaint in almost half of nasopharyngeal carcinoma (NPC) patients is a lump in the neck. Neurological symptoms occur less frequently, whereas blindness occurs in only less than one percent of cases.

Case Report: A 47-year-old male had complained about headache, nasal voice, and blindness in the right eye since a month ago. An examination found a mass in the right nasal cavity, right neck lymph nodes enlargement, damage to the right oculomotor (III), trochlear (IV), and abducens (VI) nerve, and right optic nerve (II). On funduscopy, the right macular reflex decreased. On CT scan there was a right nasopharyngeal mass extending to intracranial and right intraorbital spaces.

Conclusion: There was a direct infiltration of NPC into surrounding and intracranial structures. Unilateral blindness might be caused by expansion to middle cerebral fossa and orbita. It could also be caused by optic nerve compression from surrounding tumor tissue..

Article Info

Keywords:

Nasopharyngeal carcinoma, unilateral blindness, optic nerve.

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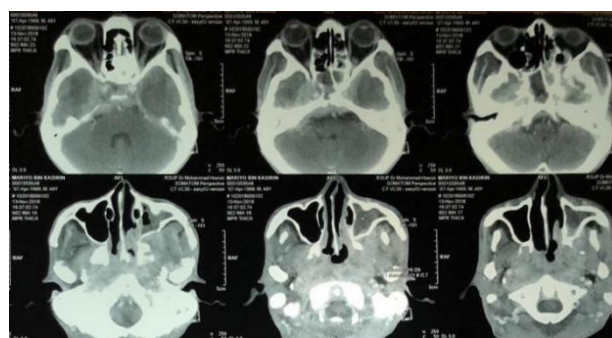
1. CASE REPORT

Nasopharyngeal carcinoma (NPC) is a type of carcinoma originating from the epithelium or mucosa and crypts that overlay the nasopharyngeal surface. Nasopharyngeal carcinoma (NPC) is a malignancy that is rare in most parts of the world and one of the most puzzling, often misdiagnosed, and difficult to understand. The incidence of NPC is low in many countries and North America, as well as Japan and India, at less than 1 per 100,000 people, but high in South China, Hong Kong, Alaska, and Greenland. The pattern of incidence shows that the prevalence of NPC is higher in Chinese natives wherever they migrate. In Southeastern Asia the overall incidence is 6.5/100,000 person-years in male and 2.6/100,000 person-years in female population. In Indonesia, NPC is the fifth largest cancer, around 5.78% of all cancers with an incidence of 6.2/100,000 population per year. The NPC in recent decades has attracted the world's attention because of the complex interactions between genetics, viruses, environmental factors, and food, which may be related to the etiology of the disease. The most common complaint is a lump in the neck in more than half of the patients. Headache occurs in more than one-third of patients. Cranial nerve disorders are usually found at an advanced stage, while visual impairment only occurs in two percent of patients [1-7].

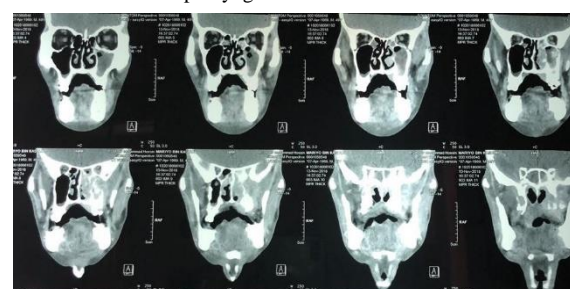
A 47-year-old man, a Sumatran tribe, came to the Emergency Unit (UGD) of RSUD Mohammad Hoesin Palembang with the main complaint of right-sided headache for one month before being admitted to the hospital. An intermittent throbbing headache that got worse over time. The medication may help temporarily, only to come back later in a few hours. The patient indicated of a lump on the right neck since 6 months ago and loss of appetite. He was also losing his right vision one month prior to admission; started with a frequent double vision which later on became more blurred, it eventually was only able to recognize the coming direction of the light. According to the patient, he ate salted fish almost every day.

On general physical examination, it was found awareness status of compost mentis (E4V5M6), adequate nutritional status, blood pressure 120/70 mmHg, pulse 82 x/minute regularly, breathing 22 x/minute and axillary temperature 36.8 °C. The eyes were proptosis, not anemic, not jaundice. From ENT examination findings obtained from endoscopic examination are a mass particularly in the right nose, enlarged lymph nodes on the right neck level II, III, IV, and V, with a size of 6x7x4 cm, well-defined, bobbing and hard on palpation. It also found enlargement of the lymph nodes in the left neck level I, and II, with a size of 2x3x2 cm, well-defined and hard on palpation. On a neurological examination, awareness of compost mentis was obtained and no sign of brain membrane excitability was found. Eye examination: anisocoric pupils, 5 mm/3 mm diameter

for direct and indirect negative light reflexes. Funduscopy of the right eye showed the well-defined papillary nerve II, cupping disc ratio 0.3, arterial/venous: 2/3, ex-macular reflex decreased, the retina was intact. There were right cranial nerves II, III, IV, and VI lesions. Normal motoric and physiological reflexes. A pathological reflex was not found. The sensory was normal. Hematological laboratory examination: mild anemia (RBC: 4.37x10⁶/μl, HGB: 11.8 g/dL, HCT: 37.6%). Blood chemistry examination: liver function, kidney function, electrolytes, and fat levels within normal limits, blood sugar level was 112 mg/L. Chest X-ray and ECG within normal limits. Coronal slices of the nasopharynx CT scan with contrast showed right-left nasopharyngeal mass extending to the left oropharyngeal parapharyngeal space of the nasal cavity, right ethmoid sinus and right maxillary sphenoid. Sphenoid/infiltration base destruction was suspected in the right-left cerebral cortex. Right-left mastoiditis was also found.



Picture 1. Nasopharyngeal CT Scan with axial cut contrast

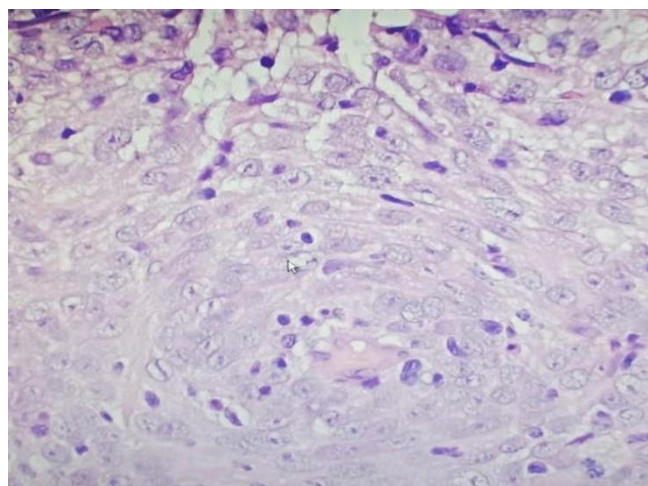


Picture 2. Nasopharyngeal CT Scan with coronal cut contrast

From the nasopharyngeal histopathology examination, a tissue preparation coated with squamous epithelium which partially dysplastic was found. Stromal tissue appeared on the subepithelium consisting of the invasive anaplastic proliferative epithelial cells, with an ovoid round nucleus, pleomorphic, irregular core membrane, partially with a prominent nucleus, narrow cytoplasm, indistinct border (syncytial), and also spindle, hyperchromatic cysts; nucleolus was not clear. These cells appeared to form a solid arrangement. Around them appeared a little necrosis and lymphocyte and PMN inflammatory cells and erythrocytes infiltration. Mitosis was difficult to find. Impression: undifferentiated non-keratinizing squamous cell carcinoma. The patient was scheduled for radiotherapy and chemotherapy.

2. DISCUSSION

This case described a male patient with nasopharyngeal carcinoma (NPC). NPC can occur at any age, however, it was found very rarely in patients under the age of 20 years and mostly in people between 45-54 years. Nasopharyngeal carcinoma is more common in men than in women with a ratio between 2-3:1. Based on the medical records in RSUP. Dr. Moh Hoesin Palembang, from January 2013 to July 2017, there were 284 cases of nasopharyngeal carcinoma. In the oncology division of the ENT department of RSMH nasopharyngeal carcinoma was the most commonly seen disease in the visiting patients.



Picture 3. Nasopharyngeal Histopathology

Although NPC can be found at all ages, rarely under 20 years, almost a quarter of patients are under thirty years old. The incidence of NPC starts to increase at the age of 20-24 years, reaches plateau between the ages of 45-54 years, then decreases. About 80% of NPC is found in productive age (30-59 years) and reaches its peak between 40-49 years old [2, 8].

Nasopharyngeal carcinoma (NPC) is a unique head-neck malignancy. Nasopharyngeal carcinoma is rarely reported in Western countries, relatively small numbers in North America, but highly frequent in South China, Hong Kong, Taiwan, Singapore, and Malaysia, which is the most common malignancy in Cantonese. The incidence pattern shows a higher prevalence of NPC in Chinese wherever they migrate. In Indonesia, NPC is the fifth largest cancer, around 5.78% of all cancers, with an incidence of 6.2/100,000 population per year. Consumption of preserved food including salted fish has been implicated in its etiology in earlier studies. Its contribution to pathogenesis of NPC remains to be determined. A decreasing trend of NPC incidence was observed in Hong Kong, Taiwan and Singapore in recent years which may be accounted by a change of dietary habits. [1, 2, 3, 5, 13].

NPC usually originates from the lateral wall of the nasopharynx, including the Rosenmuller fossa. Then it can expand into or out of the nasopharynx to the other lateral wall and/or posterosuperior to the basilar skull or base of palate, nasal cavity or oropharynx. It generally metastasizes to cervical lymph nodes. Distant metastasis can be to the bones, lungs, mediastinum and less frequently, to the liver. Cervical

lymphadenopathy is the initial symptom in many patients, and the diagnosis of NPC is often obtained from lymph node biopsy. Symptoms related to primary tumors include trismus, pain, otitis media, nasal regurgitation due to soft palate paresis, hearing loss and cranial nerve paralysis. Further growth can cause nasal obstruction or bleeding and discharge. Metastasis can cause bone pain and organ dysfunction, hence, osteoarthropathy as a paraneoplastic syndrome [7, 9].

Cranial nerve involvement was found in 22% of NPC patients at the initial examination. Cranial neuropathy occurs when the skull base is invaded by the tumor. NPC tends to spread and infiltrate the surrounding tissue and might spread superiorly involving the skull and intracranial base, and the cranial nerves. The incidence of invasion of the cranial base and brain is 12-31%. The invasion can also be anteriorly through the nasal cavity, paranasal sinuses, pterygopalatine fossa, and orbital apex. Patients with cranial nerve involvement may show ocular or orbital symptoms, such as diplopia, proptosis, and limitations of extraocular motion [7, 10].

NPC with orbital invasion is rarely reported at around 0-3%. Sympathetic innervation of the eye which ciliospinal center, is located in the lateral horn of the spinal cord from C8 to T2. Ocular and/or orbital manifestations of NPC include ocular motility disorders, blurred vision, proptosis, orbital pain, and optic disc edema. In most cases, patients experience nasal or aural symptoms at first. Most severe visual impairments occur due to tumor compression on the optic nerve. Optic atrophy usually occurs in the late stages of the disease with the symptom of severe visual acuity. NPC involving pterygopalatine and infratemporal fossa can directly affect the orbit through inferior orbital. The invasion of the orbit can be in several ways. The pterygopalatine and inferior orbital foramina are the most common routes of invasion, followed by an invasion of the paranasal sinuses. The inferior orbital fissures form a direct communication of the orbits and infratemporal fossa. The most posterior part also meets the pterygopalatine fossa, which forms direct communication between the pterygopalatine fossa and the orbital apex. Tumors in the ethmoid and/or sphenoid sinuses might erode the thin lamina papyria to reach the medial orbit and retrobulbar region, this pathway is the second most frequent pathway to the orbit. NPC rarely involves the maxillary sinus which then invades the orbital base. Orbital involvement worsens the prognosis. In stage T4 NPC, intracranial extension/cranial nerve paralysis and involvement of the orbit causes a relatively worse prognosis than the involvement of infratemporal or hypopharyngeal fossa. [7, 8, 10, 16].

Headache in NPC patients is generally associated with cranial base invasion, intracranial metastasis or cranial osteoradionecrosis, as that is found in these patients. NPC is histopathologically divided into three categories: keratinizing squamous cell carcinoma (WHO type I), nonkeratinizing squamous cell carcinoma (WHO type II), and undifferentiated carcinoma (WHO type III). This patient was diagnosed with WHO type III NPC, the most common form in Southeast Asia, including Indonesia and other areas of high incidence [8, 12].

Radiotherapy has been a standard of NPC therapy for a long time because of its anatomical location and is relatively radiosensitive, but the failure rate is high in patients who experience metastasis. The treatment of stage I NPC is radiotherapy alone. The combination of chemotherapy with radiotherapy has been widely accepted as a treatment modality for advanced NPC, but it is still not optimal. The 5-year life expectancy in stages I, II, III, and IV tumors is 95-70%, 83-65%, 76-54%, and 56-29% [2, 15].

3. CONCLUSION

The case of nasopharyngeal carcinoma has been reported with papilledema and right cranial nerve II, III, IV, VI lesions. Head CT scan with axial and coronal slices without and with contrast showed a right nasopharyngeal mass extending to the right ethmoidal sinus and sphenoid sinus accompanied by right sphenoid erosion and intracranial metastases and cerebral edema. From the nasopharyngeal biopsy, the impression is undifferentiated carcinoma. This patient is planned to undergo radiotherapy and chemotherapy.

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